

# A Case Report: Calcaneal Ewing's Sarcoma

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## Abstract

**Introduction:** Ewing sarcoma of bone represents the second most common primary malignant tumor of the bone in children and adolescents. The most common primary bony sites include the long bones, pelvis, chest wall, and spine. However, it is responsible for only 3% to 5% of cases in bones of the hands and feet.

**Case Presentation:** The case was a 13-year-old girl, who attended our clinic with a complaint of 8 months of left ankle swelling and pain. Eight months earlier, she recalled an ankle torsion for which treatment was performed with long leg splint. Her pain was better, but, her swelling remained. The X ray showed a sclerotic lesion in the calcaneus with soft-tissue mass. Next, she underwent open biopsy, and was diagnosed with Ewing's sarcoma. She underwent chemoradiotherapy and obtained below knee amputation. She was followed up at our clinic.

**Conclusions:** There are a few articles about early diagnostic effects of Ewing's sarcoma prognosis. Therefore, this study aimed at presenting an anatomical-rare case of Ewing's sarcoma as preliminary diagnosis could help the patient and lead to better outcomes.

**Keywords:** Calcaneous, Ewing Sarcoma, Malignant Tumor

## 1. Introduction

Ewing sarcoma of bone represents the second most common primary malignant tumor of bone in children and adolescents (1). Ewing sarcoma is a type of tumor that forms from a certain kind of cell in bone or soft tissue. Other names for Ewing sarcoma are a primitive neuroectodermal tumor, Askin tumor (Ewing sarcoma of the chest wall), extraosseous Ewing sarcoma (tumor growing in tissue other than bone) (2, 3). All of these names might be grouped together and called Ewing sarcoma family of tumors. The overall incidence of ESFT has remained stable over the past 25 years (4). The most common primary bony sites include the long bones (47%), pelvis (26%), chest wall (16%), and spine (6%), and a rare site is distal extremities (2). This study presented a case of calcaneal Ewing sarcoma as this tumor is responsible for only 3% to 5% of cases in the bones of the hands and feet (5). This study aimed at presenting an anatomical-rare calcaneal Ewing sarcoma with only simple ankle sprain and mismanagement of the case.

## 2. Case Presentation

A 13-year-old girl attended our clinic in Imam Khomeini hospital with a complaint of 8 months of left ankle

swelling and pain. Eight months earlier, she recalled an ankle torsion, which was treated by long leg splint. Her pain was better, yet, her swelling remained.

From 2 months ago her pain began again, and X ray was taken 2 weeks before she came to the clinic. The X ray showed a sclerotic lesion in the calcaneus with soft-tissue mass. She obtained bone-scan surveys that showed only calcaneal uptake. She came to the clinic periodically, during which her calcaneal X ray was repeated and showed increased mass size. In her laboratory data, she had anemia, elevated erythrocyte sedimentation rate (ESR), and increased lactate dehydrogenase (LDH). Therefore, she underwent computerized tomography (CT)-scan of ankle and chest, and ankle magnetic resonance imaging (MRI). Ankle CT-scan reported an ill-defined heterogeneous mass with cortical disruption and soft tissue edema. The MRI reported a signal change in calcaneal with soft tissue edema and abnormal signal in cuboid bone. The chest CT scan showed diffuse metastasis to bilateral lung. Next, she underwent open biopsy, which reported small blue round cells, while immunohistochemistry reported positive immunostaining by CD99 and Vimentin. Therefore, Ewing's sarcoma was diagnosed. She underwent chemo radiotherapy and obtained below knee amputation.

After 2 years she referred to the clinic with below knee orthosis, and was doing her daily activity independently;



Figure 1. Ankle Lat X-Ray



Figure 2. CT-Scan

follow-up was done by serial chest CT-scan to rule out lung metastasis.

### 3. Discussion

In 1921, Ewing described a tumor that was referred to as diffuse endothelioma of bone. This tumor was later renamed as Ewing's tumor (6). Ewing sarcoma is a primary malignant bone lesion usually seen in the diaphysis

of long bones and in the flat bones of young patients, in the age group of 5 to 30 years (1). Characteristically, the patient was ill, with low-grade fever, moderate leucocytosis, and anemia (7). Metastases could have occurred to lungs and to other bones. The prognosis was poor. In long bones, the tumor was seen as ill outlined diaphyseal lucencies with a surrounding soft tissue mass and periosteal reaction. There may have been osteolytic and sclerotic areas.

The differential diagnosis included infections, aneurysmal bone cyst, chondromyxoid fibroma, enchondroma, fibrous dysplasia, Ewing's tumor, osteosarcoma, Langerhan's cell histiocytosis, giant cell tumor, and chondroblastoma. Therefore, with the use of CT-scan, MRI, and open biopsy the diagnosis was confirmed.

Dahlin et al. reported 165 cases of Ewing's sarcoma, of these, only 4 cases occurred in the feet. In the case of tumors of the extremities, amputation with or without preoperative irradiation appears to be the treatment of choice, yet, the evidence is not overwhelming (7). Reinius et al. reported 12 cases of Ewing's sarcoma involving bones of hands and feet, out of a total of 377 patients. The importance of this set of patients, therefore, relates to awareness and early recognition of an unusual appearance and location of Ewing sarcoma (8).

In calcaneal Ewing's sarcoma despite the variety of methods to distinguish diagnosis, there is a long delay in diagnosis. Brotzmann et al. assessed the diagnostic delay of foot malignant tumor and stated that Ewing's sarcoma has the most delay in diagnosis in foot tumor (9). Brasme et al. claimed that delay in diagnosis of Ewing's sarcoma does not have any effect on prognosis (10). However, Jean-François Brasme in his paper said it was impossible to explain an association between a long time to diagnosis and adverse outcomes. Early diagnosis could help reduce anxiety and affliction in patients and parents, and could avoid conflicts and malpractice claims (11).

In Ewing's sarcoma, imaging plays an important role in diagnosis. In X ray, radiography lesion may be purely lytic or have variable amounts of the reactive new bone formation. Therefore, in not purely lytic lesions we must consider Ewing sarcoma as one of the differential diagnosis. Ewing's sarcoma is often associated with a lamellate or "onion skin" periosteal reaction. This appearance is caused by splitting and thickening of the cortex by tumor cells (12). The "onion-skin" appearance is sometimes followed with a "moth-eaten" or mottled appearance, and extension of soft tissue.

Computerized Tomography scan is a good imaging modality to evaluate joint extension, periosteal reaction, and a matrix of the lesion. A large soft-tissue mass is well illustrated by CT-scan, especially after intravenous administration of contrast material. Since CT scan cannot show dif-

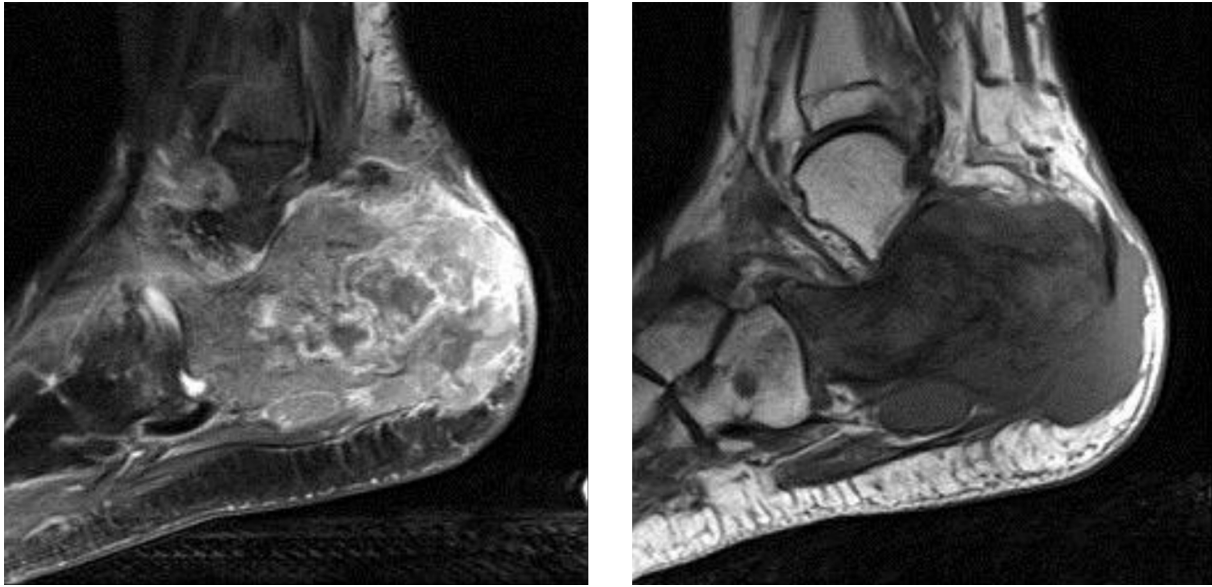


Figure 3. MRI

ferentiation of Ewing's sarcoma from other bone tumors, as well as osteomyelitis and other conditions, diagnosis may be difficult with CT scan, and false-positive results may occur (13, 14). Computerized Tomography scan is useful for assessment pulmonary involvement of metastasis, and is therefore, helpful, and must be taken for tumor staging.

In MRI, the tumor has low signal intensity on T1-weighted images compared with the normally high signal intensity of the bone marrow. On T2-weighted images, the tumor is hyper intense weighed with muscle. The importance of MRI is that is shows the soft-tissue involvement (15).

The bone scan must be done as part of staging workup and will show very "hot" lesions.

After this imaging was performed, incisional biopsy recommended achieving the pathological document.

There are a few articles about early diagnosis effect on Ewing's sarcoma prognosis. Therefore, the aim of this paper was to present an anatomical-rare case of Ewing's sarcoma as preliminary diagnosis could help the patient and lead to better outcomes.

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